Case Report

MEDICINE PALLIATIVE CARE

DOI: 10.47582/jompac.1648583

J Med Palliat Care. 2025;6(2):174-176

Desmoid-type fibromatosis in the puerperium: a case report with pelvic exenteration

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Cite this article as: Yılmaz N, Turan Bakırcı I, Çetin Arslan H, Özkan ÖF, Yeşildal C. Desmoid-type fibromatosis in the puerperium: a case report with pelvic exenteration. *J Med Palliat Care*. 2025;6(2):174-176.

Received: 28.02.2025 ◆ **Accepted**: 17.03.2025 ◆ **Published**: 23.03.2025

ABSTRACT

Desmoid-type fibromatosis (DF) is a rare, locally aggressive soft tissue tumor with no metastatic potential but a high recurrence rate. We present the case of a 28-year-old postpartum woman who developed a rapidly enlarging pelvic mass, which was later confirmed as DF via imaging and biopsy. The initial management with chemotherapy and radiotherapy was unsuccessful, necessitating radical surgical intervention with pelvic exenteration. Given its association with hormonal changes, particularly during pregnancy and the postpartum period, DF poses significant diagnostic and therapeutic challenges. This case underscores the importance of early recognition, multidisciplinary management, and individualized treatment strategies to optimize patient outcomes.

Keywords: Desmoid fibromatosis, postpartum period, pelvic exenteration

INTRODUCTION

Desmoid-type fibromatosis (DF) is an uncommon soft tissue tumor with locally invasive growth, but no potential for distant metastasis. It accounts for approximately 0.03% of all neoplasms and exhibits a predilection for young women, particularly in hormonally influenced states such as pregnancy and puerperium.[1,2] The WHO classification of DF as an intermediate tumor reflects its high propensity for local recurrence despite complete surgical excision.[3]

The etiology of DF remains incompletely understood, although hormonal factors and genetic predispositions have been implicated. It frequently arises in association with familial adenomatous polyposis (FAP), but sporadic cases also occur, with mutations in CTNNB1 leading to abnormal β-catenin accumulation.[4] This mutation contributes to uncontrolled proliferation of fibroblasts, resulting in a locally aggressive tumor with unpredictable behavior.

The management of DF remains challenging owing to its high recurrence rate and variable clinical course. Although surgical resection has historically been the standard treatment, recent evidence suggests that radical excision with negative margins does not necessarily improve long-term outcomes, prompting a shift towards conservative approaches such as active surveillance or systemic therapy.[5,6] Given its potential for local progression, DF requires a multidisciplinary approach tailored to each patient's clinical presentation and treatment response. We aim to share our experience with a case diagnosed with DF that showed rapid progression in the puerperium period and resulted in pelvic exenteration. The informed consent form was obtained from the patient.

CASE

A 28-year-old Turkish woman, gravida 3, para 1, presented with urinary retention and a palpable vaginal mass on postoperative day 4 following an elective cesarean section at 38+4 weeks of gestation. It was learned that the patient's prenatal course was uneventful. The 8x10 cm heterogeneous appearance detected in the first ultrasonographic evaluation was thought to be a hematoma or pelvic mass, and an exploratory laparotomy was performed in the clinic where she delivered, and a solid mass with deep fixation was revealed in the right pararectal region. Given its extent and uncertainty regarding its nature, the patient was referred to our tertiary center for further assessment.

Upon admission, a comprehensive evaluation was conducted, including imaging and laboratory workup. Transperineal ultrasonography demonstrated a dense solid mass measuring 11 cm in diameter, compressing the vagina. Emergency computed tomography showed a mass approximately 10 cm in diameter associated with the uterus within the pelvic

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bone borders (Figure 1). Contrast-enhanced pelvic magnetic resonance imaging revealed a large heterogeneous tumor originating from the retrorectal space, extending into the pelvic floor (Figure 2). Laboratory results, including tumor markers (CA 125, CA 19-9, CEA, AFP), were unremarkable.

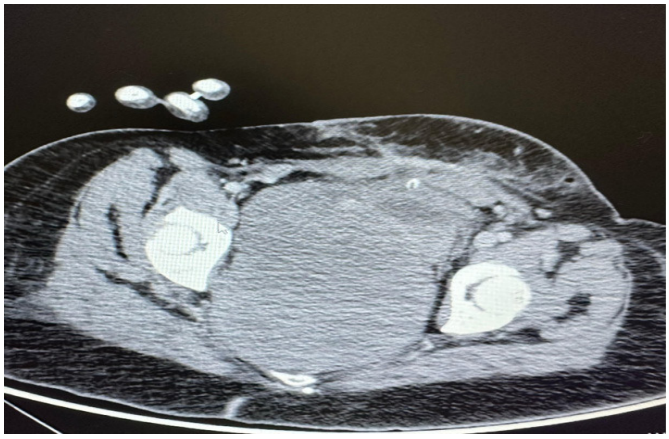

Figure 1. Heterogeneous pelvic mass with uterus within the pelvic bone borders on computed tomography

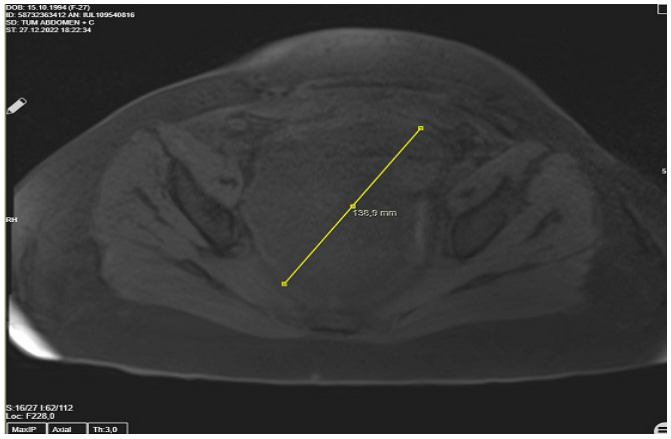

Figure 2. Magnetic resonance imaging showing a large heterogeneous pelvic mass (138×120 mm) compressing adjacent structures

A biopsy confirmed desmoid-type fibromatosis (DF) with immunohistochemical positivity for beta-catenin, vimentin, and caldesmon, raising concerns for an aggressive clinical course. The case was discussed at a multidisciplinary tumor board, and initial treatment included three cycles of chemotherapy and radiotherapy to reduce tumor burden before definitive surgery.

Despite chemotherapy, follow-up imaging showed progressive tumor growth, reaching 18x12 cm with extensive pelvic involvement. Given the tumor's aggressive nature and encroachment on adjacent structures, pelvic exenteration was deemed necessary.

On postoperative day 32, a multidisciplinary surgical team comprising general surgeons, gynecologists, and urologists performed a pelvic exenteration. A median incision was made, and upon exploration, the tumor was found infiltrating the uterus, bladder, and rectum, extending to the pelvic sidewalls. Complete en bloc resection was infeasible without significant morbidity; thus, a total abdominal hysterectomy, bilateral salpingo-oophorectomy, total cystectomy, bilateral ureterostomy, terminal ileostomy, and abdominoperineal resection were performed.

Pathological analysis confirmed extensive desmoid fibromatosis infiltration into the bladder wall, pelvic floor, and rectum, with reactive lymphadenopathy. The patient was admitted to the intensive care unit postoperatively, requiring ventilatory support. On postoperative day 4, she developed right lower limb weakness due to obturator nerve involvement. She was initiated on physiotherapy and stoma care education before discharge.

DISCUSSION

The management of desmoid fibromatosis remains a subject of ongoing debate, with treatment strategies evolving based on the emerging clinical evidence. While surgical resection has been the mainstay of therapy, studies indicate that aggressive surgery does not necessarily reduce the risk of recurrence, leading to an increased emphasis on non-surgical treatment options, such as hormonal therapy, chemotherapy, and targeted agents.[7,8] In this case, despite the initial chemotherapy and radiotherapy, tumor progression necessitated an extensive surgical approach.

The high recurrence rate associated with DF further complicates its management. Local recurrence is frequently observed even after complete excision, particularly in cases of hormone-driven growth.[4,9] The unpredictable nature of DF necessitates a case-by-case approach to balance the risks and benefits of surgery with non-invasive management strategies.

CONCLUSION

This case highlights the importance of a multidisciplinary approach in DF treatment, particularly in hormonally influenced cases, such as those occurring in the puerperium. The integration of gynecologists, oncologists, and surgical specialists allows for comprehensive decision-making, ultimately leading to favorable surgical outcomes. Further research is needed to refine treatment algorithms, incorporate molecular and genetic insights to guide individualized therapy, and improve long-term prognosis.

ETHICAL DECLARATIONS

Informed Consent

The patient signed and free and informed consent form.

Referee Evaluation Process

Externally peer-reviewed.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

Financial Disclosure

The authors declared that this study has received no financial support.

Author Contributions

All of the authors declare that they have all participated in the design, execution, and analysis of the paper, and that they have approved the final version.

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